

## Synovial Sarcoma of the Abdominal Wall

Jeanine McNeill, M.D., and Y. Vien Nguyen, B.S.

We present the case of a 43-year-old man who presented with metastatic abdominal wall synovial sarcoma. CT of the abdomen showed a 6 cm anterior abdominal wall mass with a nodular, non-homogeneous pattern of enhancement. A chest radiograph showed metastases to the lungs.

### Introduction

Synovial sarcoma represents a relatively common type of primary soft tissue malignancy. Despite the name, the tumor rarely arises from an intra-articular location, although it usually arises in an extremity or limb girdle. Rarely, synovial sarcoma may in unexpected locations such as the abdominal wall. We report the imaging findings of a patient who presented with metastatic abdominal wall synovial sarcoma.

### Case Report

A 43-year-old African American male was diagnosed in an emergency room with persistent hemoptysis. The only significant aspect of his past history was a pulled muscle injury in his right chest wall 10 days previously during a

routine workout. He had no other sites of pain or known masses. Initial chest radiograph showed multiple large masses throughout both lungs, which were confirmed with CT (Figs. 1-2). Physical exam revealed a 4-5 cm mass in the right lower quadrant anterior abdominal wall, which appeared to be deep to the subcutaneous fat and was non-tender to palpation. CT of the abdomen confirmed the presence of a 6 cm anterior abdominal wall superficial mass (Fig 3-4). An incisional biopsy was performed showing the mass to be directly deep to the external oblique fascia, and circumscribed by a shell or rim. Microscopic examination showed a highly cellular spindle cell neoplasm, consisting of mostly ovoid to spindle-shaped cells with high nuclear-cytoplasmic ratio in sheets or fascicles. The nuclei were pleomorphic and hyperchromatic with frequent mitotic figures. A single minute focus of epithelioid columnar cells forming glandular structures was present on the background of spindle cells. Immunohistochemical markers showed strong positivity for viable epithelioid cells with CAM5.2, AE1/AE3, and EMA. In situ hybridization confirmed the translocation of chromosome t(X,18) and a final diagnosis of biphasic synovial sarcoma of the abdomen was made.

### Discussion

Synovial sarcoma represents a relatively common type of primary soft-tissue malignancy accounting for 2.5-10.5% of all primary malignant soft-tissue neoplasms [1]. Ab-

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Abbreviations: CT, computed tomography

Jeanine McNeill, M.D. (Email: jamcneill@yahoo.com) is with Renaissance Radiology, Riverside, CA 92503, United States of America.

Y. Vien Nguyen, B.S., is at the Western University of Health Sciences, Pomona, CA 91766-1854, United States of America.

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dominal wall synovial sarcomas are quite rare with only 44 cases having been reported in the literature between 1950 and 2005 [2]. Despite the nomenclature, these tumors rarely arise from intra-articular locations. The most common location of origin is near the knee in the popliteal fossa. The tumor occurs most frequently in adolescents and young adults, with the majority of the patients presenting at 15-40 years of age [3]. No significant gender preponderance has been documented. No race or ethnic predilection has been reported [4]. However, recent cytogenetic studies found a highly specific association between synovial sarcoma and chromosome t(X;18) translocation and SYT-SSX gene fusion products. Greater than 90% of synovial sarcomas express this chromosomal abnormality, with 67% and 33% of tumors possessing SYT-SSX1 and SYT-SSX2 gene fusion types, respectively. The prognostic difference between these two gene fusion types remains controversial [2].

Most patients with synovial sarcoma present with a palpable soft tissue mass or swelling associated with pain or tenderness and possible minor limitation to range of motion, often slow-growing initially. Constitutional symptoms such as weight loss are rare [5]. Abdominal synovial sarcomas are associated with vague intestinal symptoms, abdominal pain or mass, and most reach a large size prior to clinical presentation. These tumors usually occur equally on both sides of the abdomen, and are more common in the lower half of the abdomen [5]. Metastases are present

in 16-25% of patients at their initial presentation with the most frequent metastatic site being the lung as in this case's presentation [6].

Synovial sarcoma does not arise from intra-articular tissue, as the name would suggest. To date, the pathogenesis of the synovial sarcoma remains unresolved. The exact line of differentiation remains a mystery and several theories exist: (1) normal non-neoplastic synovium of joints, tendon sheaths, or bursae; (2) specialized forms of mesenchymal tissue (so-called arthro-genous mesenchyme); or (3) primitive mesenchyme or ordinary fibrous connective tissue analogous to the synovial membrane formation in traumatic or occupational bursae (synovial metaplasia). However, findings on histochemical and ultrastructural studies have persuaded some reviewers to believe that synovial sarcomas arise from primitive mesenchyme, rather than preformed synovial cells [5].

There are three main histologic subtypes of synovial sarcoma: biphasic, monophasic and poorly differentiated [7]. The biphasic type represents 20-30% of lesions and has both mesenchymal spindle cell components and an obvious epithelial component usually forming glands. The monophasic type is the most common (50%-60%), in which the spindle cell component predominates. Poorly differentiated synovial sarcomas are epithelioid in morphology and have high mitotic activity. This type has the poorest prognosis [5]. Cytogenetic studies have found that biphasic tumors predominantly express SYT-SSX1



Figure 1A. 43-year-old man with metastatic synovial sarcoma of the abdominal wall. A, Chest radiograph shows bilateral lung masses.

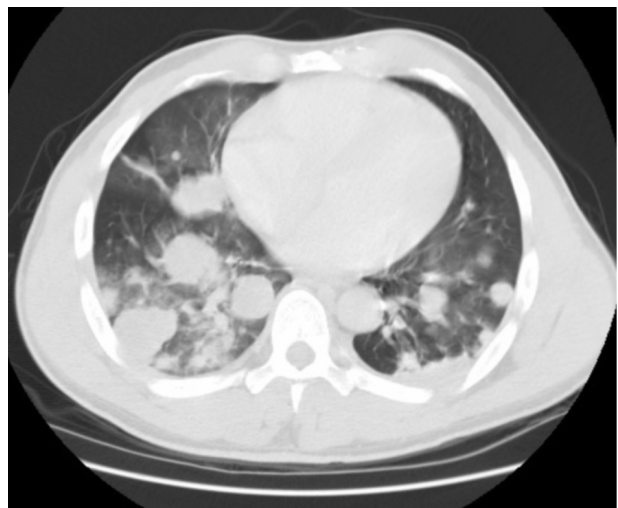


Figure 1B. B, CT through lung bases shows the masses are not cavitated.

fusion transcription, while monophasic tumors express either SYT-SSX1 or SYT-SSX2 genetic rearrangement [1]. However, some studies have shown that biphasic synovial sarcoma arising in the abdomen can have SYT-SSX2 gene rearrangement [8]. In addition to microscopic evaluation of these tumors, a recent immunohistochemical profile suggests that EMA, cytokeratin AE1/AE3 and E-cadherin, in combination with CD34 negativity, are the most useful and sensitive markers for diagnosing synovial sarcoma [2].

Radiographs appear normal in approximately 50% of cases [9]. Synovial sarcoma predominantly present on x-ray films as round or oval, lobulated swellings or masses of moderate density, usually located in close association to large joints and usually without involvement of the underlying bone. In cases with associated bone erosion, often an indolent non-aggressive appearance is reported. Aggressive bone invasion and destruction is less common, occurring in approximately 5% [9]. Calcification is present in up to 30% of cases on radiography. These are usually eccentric or peripheral within the soft-tissue mass, represented by multiple small spotty radio-opacities. Extensive calcification is associated with an improved prognosis. Computed tomography usually presents with a heterogeneous soft tissue mass with attenuation similar or slightly less than that of muscle. Areas of necrosis or hemorrhage presenting as lower density areas are common. Contrast enhanced CT

shows heterogeneous enhancement in 89%-100% of cases [10]. Nodular enhancement may also be seen. MRI is the optimal radiological modality for assessing the extent and characteristics of synovial sarcomas. Intermixed areas of low, intermediate and high signal intensity on long repetition time images has been marked as the triple sign presumably resulting in a mixture of solid cellular elements, hemorrhage or necrosis and calcified or fibrotic regions, with hemorrhage, fluid levels, and septa creating the bowl of grapes sign. This triple sign has been described as occurring in 35-57% [11]. However, the triple sign is also seen in other soft tissue tumors; therefore, this finding alone lacks a high degree of specificity. Angiographic studies of synovial sarcoma frequently reveal a prominent vascularity of primary lesions and metastatic disease [5].

As with most soft tissue tumors, the treatment of choice is wide excision followed by adjuvant chemotherapy and radiation. The 5-year survival rate for intermediate to high-grade sarcoma is 36%-76% [12]. The clinical course of synovial sarcoma is characterized by a high rate of local recurrence (30%-50%) and metastatic disease. Metastases are present in 16%-25% of patients at their initial presentation, with the lung being the most frequent site of metastasis (94% of cases) [3]. Less commonly, metastasis can also occur in the lymph nodes (4%-18%) and bone (8%-11%). The majority of metastases occur within the



Figure 1C. Axial CT of abdomen with oral but not intravenous contrast shows mixed density soft tissue mass arising within the right external oblique muscle. There is no invasion into the abdominal cavity.



Figure 1D. Post-contrast CT shows a nodular, non-homogeneous pattern of enhancement.

first 2-5 years after treatment [5]. In addition, it has also been suggested that SYT-SSX2 gene fusion, monophasic synovial sarcoma, size less than 5 cm, less than 15 years of age, and distal extremity location have better prognostic outcomes, although there is still an on-going debate in regard to the significance of these findings [13].

Our case presentation shows an atypical synovial sarcoma location within the abdominal wall with commonly seen metastatic disease to the lungs. Synovial sarcoma is essential in a differential diagnosis if multiple masses are seen in the lungs with the characteristic “cannon ball” appearance. Since metastases of this nature are usually hematogenous, the following differential diagnosis should be considered: (1) choriocarcinoma, (2) head/neck cancer, (3) renal cell cancer, (4) colon cancer, and (5) sarcomas. Although rare, proper diagnosis of synovial sarcoma can expedite treatment and management of the disease and improve prognostic outcomes.

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